abcam

Product datasheet

Recombinant human Noggin protein ab134390

Description

Product name	Recombinant human Noggin protein
Biological activity	Determined by its ability to inhibit 5.0 ng/ml of BMP4 induced alkaline phosphatase production by ATDC-5 chondrogenic cells. The expected ED_{50} for this effect is 2.0-3.0 ng/ml of Noggin.
Purity	> 95 % SDS-PAGE. Purity is greater than 95% by SDS-PAGE gel and HPLC analyses.
Expression system	HEK 293 cells
Accession	Q13253
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Sequence	QHYLHIRPAP SDNLPLVDLI EHPDPIFDPK EKDLNETLLR SLLGGHYDPG FMATSPPEDR PGGGGGAAGG AEDLAELDQL LRQRPSGAMP SEIKGLEFSE GLAQGKKQRL SKKLRRKLQM WLWSQTFCPV LYAWNDLGSR FWPRYVKVGS CFSKRSCSVP EGMVCKPSKS VHLTVLRWRC QRRGGQRCGW IPIQYPIISE CKCSC
Predicted molecular weight	23 kDa
Amino acids	28 to 232

Specifications

Our Abpromise guarantee covers the use of **ab134390** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Applications	SDS-PAGE HPLC Functional Studies
Form	Lyophilized

Preparation and Storage

Stability and Storage	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles. This product is an active protein and may elicit a biological response in vivo, handle with caution.
Reconstitution	Centrifuge vial prior to opening. Reconstitute in water to 0.1-1.0 mg/ml. Do not vortex. Store at 2°C to 8°C for 1 week, or prepare for extended storage. If the product is required to be stored after reconstitution it must be prepared for extended storage. Follow reconstitution with further dilution in a buffer containing a carrier protein (example 0.1% BSA). Store working aliquots at -20°C to -80°C. Avoid repeated freeze-thaw cycles. The product can then be stored for 3 months.

General Info

Function	Essential for cartilage morphogenesis and joint formation. Inhibitor of bone morphogenetic proteins (BMP) signaling which is required for growth and patterning of the neural tube and somite.
Involvement in disease	Defects in NOG are a cause of symphalangism proximal syndrome (SYM1) [MIM:185800]. SYM1 is characterized by the hereditary absence of the proximal interphalangeal (PIP) joints (Cushing symphalangism). Severity of PIP joint involvement diminishes towards the radial side. Distal interphalangeal joints are less frequently involved and metacarpophalangeal joints are rarely affected whereas carpal bone malformation and fusion are common. In the lower extremities, tarsal bone coalition is common. Conducive hearing loss is seen and is due to fusion of the stapes to the petrous part of the temporal bone. Defects in NOG are the cause of multiple synostoses syndrome type 1 (SYNS1) [MIM:186500]; also known as synostoses, multiple, with brachydactyly/symphalangism-brachydactyly syndrome. SYNS1 is characterized by tubular-shaped (hemicylindrical) nose with lack of alar flare, otosclerotic deafness, and multiple progressive joint fusions commencing in the hand. The joint fusions are progressive, commencing in the fifth proximal interphalangeal joint in early childhood (or at birth in some individuals) and progressing in an ulnar-to-radial and proximal-to-distal direction. With increasing age, ankylosis of other joints, including the cervical vertebrae, hips, and humeroradial joints, develop. Defects in NOG are the cause of tarsal-carpal coalition syndrome (TCC) [MIM:186570]. TCC is an autosomal dominant disorder characterized by fusion of the carpals, tarsals and phalanges, short first metacarpals causing brachydactyly, and humeroradial fusion. TCC is allelic to SYM1, and different mutations in NOG can result in either TCC or SYM1 in different families. Defects in NOG are a cause of stapes ankylosis with broad thumb and toes (SABTS) [MIM:184460]; also known as Teunissen-Cremers syndrome. SABTS is a congenital autosomal dominant disorder that includes hyperopia, a hemicylindrical nose, broad thumbs, great toes, and other minor skeletal anomalies but lacked carpal and tarsal fusion and symphalangism. Defects in NOG are the cause of brachydactyly type B2 (BDB2) [MIM:611377]. BDB2 is a subtype of brachydactyly characterized by hypoplasia/aplasia of distal phalanges in combination with distal symphalangism, fusion of carpal/tarsal bones, and partial cutaneous syndactyly.
Sequence similarities	Belongs to the noggin family.
Cellular localization	Secreted.

Our Abpromise to you: Quality guaranteed and expert technical support

- Replacement or refund for products not performing as stated on the datasheet

- Valid for 12 months from date of delivery
- Response to your inquiry within 24 hours
- We provide support in Chinese, English, French, German, Japanese and Spanish
- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit https://www.abcam.com/abpromise or contact our technical team.

Terms and conditions

- Guarantee only valid for products bought direct from Abcam or one of our authorized distributors